International Journal of Research In Pharmacovigilance and Pharmacotherapeutics

Longitudinal extensive transverse myelitis-a case report

Varadaraju Gurupriya*[1], S. Divya[1], Robin George[2], E. Sunil Kumar[2]

[1]Pharm D intern, Department of Pharmacy Practice, Seven Hills College of Pharmacy, Tirupati, Andhra Pradesh, India.
[2]Department of Pharmacy Practice, Seven Hills College of Pharmacy, Tirupati, Andhra Pradesh, India.

ABSTRACT

Longitudinal Extensive Transverse Myelitis (LETM) characterized by spinal cord inflammation extending vertically through three or more vertebral segments. Early serological testing for NMO-IgG antibodies in suspected LETM is important for prompt diagnosis, indicating prognosis and establishing appropriate therapy to reduce the risk of future relapses or disease progression. This is a case of 35-year-old female complaining of weakness of both lower limbs, bladder and bowel disturbances with curdle like sensation, loss of sensation. MRI C and D spine plain and contrast showed T2 hyperintensity in spinal from C4 to D7 s/o of demyelination, thinning of spinal cord noted at upper dorsal level. Anti nuclear antibody ELISA test revealed positive indicating the presence of NMO IgG antibodies. Due to poor prognosis of the disease patient was advised to be in observation for 15 days and planned for 2 doses of cyclophosphamide 750 mg. Mean while symptomatic treatment was also given. Performing NMO antibodies test as early as possible in the case of symptomatic assessment of either LETM or Multiple sclerosis, which facilitates the choice of treatment for better patient care and enhanced health related quality of life.

Keywords: Cyclophosphamide; LETM; Multiple sclerosis.

ISSN: 2582-0672
Research Article

Corresponding Author

Name: Dr. E Sunil Kumar
Email: sunilkumarellampati@gmail.com

Article Info

Received on: 15-11-2019
Revised on: 08-12-2019
Accepted on: 20-12-2019

DOI: https://doi.org/10.33974/ijrpp.v1i1.160

INTRODUCTION

Neuromyelitis optica spectrum disorders (NMOSD)or Neuro myelitis optica (NMO) are inflammatory disorders of central nervous system characterized by severe, immune mediated demyelination and axonal damage predominantly targeting optic nerves and spinal cord.[1] NMOSD include either Longitudinal Extensive Transverse Myelitis (LETM) or Optic neuritis.

LETM is characterized by spinal cord inflammation extending vertically through three or more vertebral segments. It may occur as an uncommon manifestation of SLE or other autoimmune diseases[2]. Signs and symptoms include neurological dysfunction in motor and sensory tracts on both sides of spinal cord. Involvement of motor and sensory control pathways frequently produce altered sensation, Weakness, Urinary or bowel dysfunction[3].

Early serological testing for NMO-IgG antibodies in suspected LETM is important for prompt diagnosis, indicating prognosis and establishing appropriate therapy to reduce the risk of future relapses or disease progression. Patients with NMO-IgG seropositivity are considered at increased risk of future relapses and development of whole NMO disease spectrum. Other diagnostic tests include MRI spine, Neurophysiological tests. [4]

Table 1: NMO-Antibody test

NMO-NMOSD-SERUM
Neuro myelitis optica IgG antibodies STRONGLY POSITIVE Anti-myelin oligodendrocyte glycoprotein (MOG)
IgG antibodies NEGATIVE

Table 2: Complete blood count

Lab Parameter	Observed Value	Normal Value
Haemoglobin	11.5g/dl	12-16g/dl
Red blood cells	4.03mill./cumm	4.5-5mill./cumm
White blood cells	11900cells/cumm	4000-11000cells/cumm
Platelets	1.68 L/cumm	1.5-4 L/cumm
Erythrocyte sedimentation rate	14 mm/1st Hr	20-40mm/1st Hr

Table 3: Pharmacological treatment provided

S.no	Dosage form	Medications	Dose	Route	Frequency
1	Tablet	Gabapentin + Nortryptylline	100mg	Oral	TID
2	Tablet	Shelcal (calcium)	500mg	Oral	OD
3	Injection	Pantoprazole	40mg	Intravenous	TID
4	Injection	Methyl prednisones	1g	Intravenous	OD
5	Injection	Cefoperazone + Sulbactum	1.5g	Intravenous	BD
6	Capsule	Vitamin D	60,000u	Oral	Weekly once
7	Injection	Cyclophosphamide	750mg	Intravenous	Monthly once

The goal of treating acute events is to improve relapse symptoms and restore neurological function[5]. Yanagawa et al. reported that early detection of NMO-IgG combined with proper immunotherapy represents a key to a good, long term prognosis[6]. The rationale for treatment of acute and recurrent attacks is based upon evidence that humoral immunity plays a role in pathogenesis of NMOSD. Intial treatment with high dose intravenous methylprednisolone 1g daily for three to five consecutive days. For patients with severe symptoms, unresponsive to glucocorticoids, Therapeutic plasma exchange is the suggested rescue treatment. Patients with severe attacks do better if plasma exchange is started early as adjunctive therapy with glucocorticoids. If both immunotherapy and plasma exchange do not improve symptoms, treatment with intravenous immunoglobulins or an escalation to cytoablative therapy such as cyclophosphamide may be considered. [5]

CASE STUDY

We present the case of a 35-year-old female (K/C/O paraparesis 2016 recovered) complaining of weakness of both lower limbs, bladder and bowel disturbances with curdle like sensation, loss of sensation below chest area since 2 months. Patient also presented with history of chronic backache from 2 years, 2 months back she developed paresthesias ascending type in both lower limbs within 3 days up to chest area, unable to walk or move around in bed. On examination patients blood pressure was 130/90 mm of Hg, pulse rate was 70 bpm, power of upper limbs 4/5, lower limbs 1/5. Gait could not be tested.

Serum electrolytes, Renal function tests and liver function tests were found to be normal. MRI C and D spine plain and contrast showed T_2 hyper intensity in spinal from C_4 to D_7 s/o of demyelination, thinning of spinal cord noted at upper dorsal level. Anti-nuclear antibody ELISA test revealed positive indicating the presence of antibodies.

Serum analysis for the presence of NMO antibodies showed in (Table 1)

Patient found to have elevated levels of plasma lactate-53mg/dl (4.50-19.80). With the evidence of laboratory investigations, patient was diagnosed with "NMO antibodies associated longitudinal extensive transverse myelitis". Treatment include patient initially treated with 1g of intravenous methylprednisonolone, no significant improvement in the patient. 5 cycles of plasmapheresis, also known as plasma exchange (PLEX) were done, which is often recommended for moderate to severe forms of LETM. Due to poor prognosis of the disease patient was advised to be in observation for 15 days and planned for 2 doses of cyclophosphamide 750 mg. Meanwhile symptomatic treatment was also given.

After giving 2 doses of cyclophosphamide 750 mg. Patient was stabilized and symptoms were relieved.so patient was discharged with the medications:Gabapin NT 400/10 twice daily, pantoprazole 40 mg once daily before breakfast, calcium and multivitamin twice daily and monthly cyclophosphamide during follow up to reduce the number of relapses.

DISCUSSION

LETM is an inflammatory condition of the spinal cord causing variable levels of morbidity and mortality[4]. Traditionally the majority of LETM cases were thought to be characterized by perivascular infiltration by monocytes and lymphocytes in the spinal cord lesion. To distinguish LETM from multiple sclerosis and to provide appropriate therapy we additionally emphasize the clinical importance of NMO-IgG serological status. A previous report has suggested that NMO–IgG antibodies are present in approximately 38% of LETM patients following an initial attack and is associated with greater than 60% risk of relapse within 1 year and development of optic neuritis[7]. Encouragingly, LETM is often responsive to acute immunotherapy with steroids, plasma exchange[8]. We observed that in this case the patient was not responded

well to the steroid therapy and plasmaphresis, so an alkylating agent such as cyclophosphamide was considered.

CONCLUSION

LETM can be specifically diagnosed with the help of NMO-IgG antibodies test, so that we can differentiate LETM with Multiple sclerosis in initial stages. We suggest to perform NMO antibodies test as early as possible in the case of symptomatic assessment of either LETM or Multiple sclerosis, which facilitates the choice of treatment for the better patient care and enhanced health related quality of life. Further studies with larger cohort should be needed to consolidate the findings and it potentially leads to therapeutic recommendations in majority of the NMO-IgG seropositive patients.

ABBREVATIONS

NMOSD: Neuromyelitis optica spectrum disorders, LETM: Longitudinal extensive transverse myelitis, PLEX: Plasma exchange.

ACKNOWLEDGEMENTS

We acknowledge management of the Sri Venkateswara Institute of Medical Sciences& Sri Padmavati Medical College Hospital For Women ,Tirupati, Andhra Pradesh, India for their valuable support.

REFERENCE

1. Glisson, C. C. (2016). Neuromyelitis optica spectrum disorders.
2. Evangelopoulos E M, Koutsis G., Neuromyelitis optica spectrum disease with positive autoimmune indices: A case report and review of the literature, Case reports in medicine, volume 2011, http://dx.doi.org/10.1155/2011/393568.
3. John Hopkins Medicine Neurology and neurosurgery(https://www.hopkinsmedicine.org/neurology_neurosurgery/centers_clinics/project_restore/conditions/transverse-myelitis.html)
4. Nightingale H,With Erick J,Wilkins A,Diagnosis of longitudinal extensive transverse myelitis,BMJ Case Rep. 2011; 2011: bcr1020103444 doi: 10.1136/bcr.10.2010.3444
5. Jarius S, Wildemann B. [Neuromyelitis optica]. Nervenarzt 2007; 78: 1365– 1377.
6. Yanagawa K,Kawachi I, Toyoshima et al., "Pathologic and immunologic profiles of a limited form of neuromyelitis optica with myelitis," Neurology, vol. 73, no. 20, pp. 1628–1637, 2009.
7. Weinshenker BG, Wingerchuk DM, Vukusic S, et al. "Neuromyelitis optica IgG predicts relapse after longitudinally extensive transverse myelitis". Ann Neurol 2006;59:566–9
8. Wingerchuk DM, Lennon VA, Lucchinetti CF, Pittock SJ, Weinshenker BG. The spectrum of neuromyelitis optica. Lancet Neurol 2007; 6: 805–15.